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CLINICAL STUDY

Clinical spectrum and surgical outcomes in spherophakia: a prospective interventional study

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Abstract

Purpose **To study the varied clinical presentations of patients with spherophakia, their management using surgical methods, and the clinical outcomes.**

Patients and methods **A prospective interventional study of 13 patients of spherophakia who presented to us from January 2014 and were followed up over the course of their treatment, and the data were documented for analysis.**

Results **In all, 26 eyes of 13 patients were reviewed and the median age of presentation was 12 ± 12.05 years. All patients had a bilateral presentation with 22 eyes having lenticular myopia with a mean refractive error of −11.5 ± 12.945 DS. Ten eyes presented with glaucoma of which six had raised intraocular pressure (IOP) >21 mm Hg. A total of 23 eyes underwent lens extraction for dislocation/subluxation. Lens extraction helped lower overall IOP. Refractive rehabilitation was done with ACIOL, posterior chamber intraocular lens (PCIOL) with capsular tension ring, and scleral-fixated intraocular lens (SFIOL) in respective cases with ACIOLs being the most commonly used option.**

Conclusions **Spherophakia is a rare condition, which exhibits a varying degree of lenticular myopia, glaucoma, and subluxation of the crystalline lens. Lensectomy with proper rehabilitation using ACIOL, PCIOL, or SFIOL is a method of managing subluxation and unacceptable myopia. Lensectomy may also be a viable option of controlling glaucoma alongside medications and glaucoma surgery for the management of glaucoma in such cases.**

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Introduction

Spherophakia is a rare congenital bilateral eye disorder, which presents with weak zonules around a smaller and more spherical crystalline lens with an increased anteroposterior thickness of the lens, and highly myopic eye.[1] The lens zonules are developmentally hypoplastic and abnormally weak and due to non-attachment of the posterior zonules to the equatorial zone of the lens, the lens changes its normal shape to spherical. The lens may undergo subluxation or dislocation from the patellar fossa, leading to defective accommodation. The disease can present as an isolated condition or may run in families[2] and such cases have been reported in multiple lineage studies. Peculiarly in all these families, consanguinity seemed to be an important finding.[1,3] Subluxation of lens may occur anteriorly, inferiorly, or posteriorly,[4] and may lead to pupillary block glaucoma.[5] We hereby describe an interventional study of spherophakia patients who presented to our center with different manifestations of the disease and have been managed accordingly.

Materials and methods

This prospective interventional study evaluated the management strategies and outcomes of different presentations of microspherophakia cases, which were referred to our center. The study was done adhering to the tenets of the Declaration of Helsinki after taking proper approval from the Institutional Review Board of our hospital. Written informed consent was obtained from all included candidates.

Included in this study were 13 patients of microspherophakia who presented to our center from January 2014 onwards and all included patients were followed up for a minimum period

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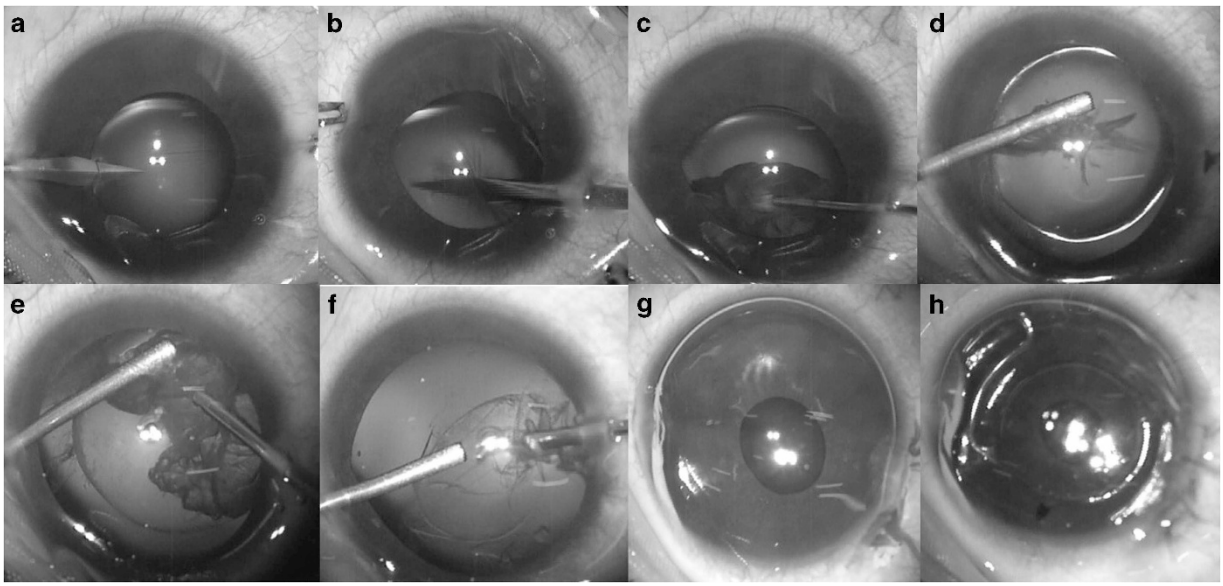

Figure 1 Endocapsular lens aspiration with insertion of ACIOL. (a) MVR entry is made in the cornea and extended to make a nick in the anterior capsule of the lens. (b) Two nicks are made in the anterior capsule with MVR blade. (c) Adrenaline is injected through one of the nicks for hydro dissection. (d) Irrigation cannula through opening and bag stabilized. (e) 25G vitrectomy cutter is inserted through one of the nicks and lens aspiration done in I–A mode. (f) Capsular bag is eaten and limited anterior vitrectomy is done with the cutter in cut I–A mode. (g) Pilocarpine followed by air is injected intracamerally and the iris is stroked with a Sinkey hook to constrict the pupil. (h) One of the corneal incisions is enlarged for the insertion of ACIOL and a PI is done. A full colour version of this figure is available at the *Eye* journal online.

of 1 year till November 2016. Data regarding demography, age at presentation, chief complaints, physical and ophthalmic examination, intraoperative findings, and postoperative examination were recorded, with a review of medical records and medication history. All patients were asked for family history status of a similar illness. All patients were evaluated for systemic status by a general physician/pediatrician.

Complete ophthalmic examination included preoperative best-corrected visual acuity (BCVA) using Snellen's chart at 6 m, intraocular pressure (IOP) using non-contact applanation tonometer (NT-510, Nidek, Japan), slit lamp examination for anterior segment evaluation, dilated binocular indirect ophthalmoscopy for fundus, retinoscopy (phakic and aphakic) for refractive status, non-contact specular microscopy (SP 3000P, Topcon, Oakland, CA, USA) to determine the endothelial cell count (ECD), white-to-white (WTW) measurement using Orbscan II (Orbtek, Salt Lake City, UT, USA), central corneal thickness measurement with ultrasound pachymeter (IOPac Advanced, Heidelberg Engineering GmbH, Germany), and biometry using IOLMaster 500 (Carl Zeiss Meditec, Jena, Germany). Patients with poor glow underwent a B-scan ultrasonography. Ultrasound biomicroscopy (Sonomed, Lake Success, NY, USA) with 50 Hz probe was performed for all cases to further delineate the anterior segment details. The patients also underwent screening for urine homocysteine levels and serum homocysteine levels were measured for those in whom urine screening came positive. Postoperatively, BCVA, IOP, refractive status, fundus, and endothelial counts were checked for all.

Surgical techniques

All surgeries were performed under sterile operation settings by the same surgeon (SK). General anesthesia was used for patients <16 years of age and peribulbar anesthesia for those above (Supplementary Video 1). Two clear corneal incisions were made using a 23G microvitreoretinal blade (MVR; Alcon Laboratories, Inc., Fort Worth, TX, USA) at 10 o'clock and 2 o'clock positions with further insertion of the tip of the blade into the anterior capsule of the lens (Figure 1a and b). A visco-dispersive agent (Viscoat; Alcon Laboratories, Inc.) was injected at the site of the zonular stretch to prevent vitreous entry into the anterior chamber. Vitrectomy cutter of Centurion system (25G, Alcon Laboratories, Inc.) was introduced through one incision and irrigation cannula through the other within the capsular bag and keeping the cutter in irrigation–aspiration cut (I–A cut) mode (Figure 1c and d), the lens matter was aspirated keeping vacuum at 400 mm Hg, aspiration flow rate at 50 c.c.s./min and cut rate at 100 c.p.m. (Figure 1e). The capsular bag was then eaten keeping the cutter vacuum at 250 mm Hg, aspiration flow rate at 20 c.c.s./min and cut

rate at 4000 c.p.m. and a limited anterior vitrectomy was performed in cut I–A mode to free the anterior chamber of vitreous strands (Figure 1f). An anterior chamber intraocular lens (IOL) was placed, after enlarging the main entry wound using a 5.2 mm single-bevel keratome (Alcon Laboratories, Inc.) (Figure 1g and h). The anterior chamber was formed by sterile air, and steroids–antibiotics combination was injected subconjunctivally. A peripheral iridotomy (PI) was made superiorly with the help of vitrectomy cutter in I–A cut mode in cases where ACIOL was implanted. The main corneal wound and the two side ports were sealed with stromal hydration using balanced salt solution and 10-0 nylon suture was placed if necessary. In cases where bag needed to be preserved, after making MVR entries, capsulorhexis was started from the 12 o'clock meridian with a 26-gauge cystitome under sodium hyaluronate 1.4% (Healon GV, Pharmacia Upjohn, Kalamazoo, MI, USA) and finished with Utrata capsulorhexis forceps in the anticlockwise direction to avoid excessive zonular stretching. A modified capsular tension ring (M-CTR) was placed to stabilize the bag. Viscoelastic was removed from the anterior chamber with I–A probe. In all cases, a superior clear corneal incision was made with a 2.2 mm single-bevel keratome (Alcon Laboratories, Inc.) (not made in patients finally left aphakic). Healon 1.0% (Pharmacia Upjohn) was injected to stabilize the anterior chamber and repeating the A-scan biometry under sterile conditions, a posterior chamber IOL was implanted if the bag was intact. In eyes with a posteriorly dislocated lens, a core vitrectomy with 25G+ vitrectomy cutter was performed followed by fragmentation of dropped cataractous spherophakia lens with fragmatome, with a scleral-fixated lens insertion and haptic burial under the scleral tunnel.

Postoperatively, the patients were advised topical prednisolone phosphate 1% (P-lone, Syntho Pharmaceuticals Pvt. Ltd., Lucknow, India) six times a day, moxifloxacin hydrochloride 0.5% three times a day (Vigamox, Alcon Laboratories, Inc.), and homatropine 2% four times a day. Slit lamp examination was done postoperatively to assess the corneal clarity, the anterior chamber integrity, and the status of the IOL. IOP was measured using non-contact applanation tonometer. Specular microscopy was done at 1 month postoperatively to determine the endothelial cell status.

Results

Age of patients at presentation ranged from 2 to 40 years with a median age of 12 ± 12.06 years. Six patients were 10 years or younger and seven patients were older. All the patients were followed up for a minimum period of 1 year. Clinical details of the included patients have been tabulated in Table 1 along with the intraoperative and postoperative findings. The commonest presenting complaint was the diminution of vision ($n = 24$) followed by pain ($n = 5$) with 22 eyes having high myopia. Other patients were effectively aphakic due to the dislocation/significant subluxation of the crystalline lens from the patellar fossa. All the examined patients had a bilateral presentation of the disease and 20 eyes presented with some form of subluxation of the spherical lens, either anteriorly, superonasally, or inferiorly. In all, 19 eyes had shallow or irregular anterior chamber with 8 eyes displaying iridodonesis and 6 eyes with frank phacodonesis. Six eyes presented with corneal edema, the most common cause being shallow anterior chamber and corneo-lenticular touch ($n = 4$) followed by raised IOP ($n = 2$).

Snellen's visual acuity was determined for 26 eyes and a BCVA of $>6/60$ was present in 13 eyes. The mean refractive error at presentation was -11.5 ± 12.45 DS. Mean axial length of 24 eyes was 22.86 ± 2.13 mm with mean WTW of 12.43 ± 0.85 mm and mean keratometry value of 43.2 ± 2.17 mm. Six eyes had axial lengths of >24 mm. The ocular parameters have been tabulated in Table 2.

In all, 23 eyes were operated for lensectomy in view of the crystalline lens in the anterior chamber, an anteriorly dislocated lens with or without pupillary block glaucoma, inferior dislocation of the lens causing lenticular astigmatism, or posteriorly dislocated lens in the vitreous cavity. Refractive rehabilitation was done using anterior chamber IOLs in 13 eyes, posterior chamber IOL with CTR fixation in 3 eyes, and scleral-fixated glued IOL in 2 eyes. Seven eyes were left aphakic. BCVA after surgery was $>6/60$ in 18 (78.3%) operated eyes with $>6/18$ in 10 (43.5%) eyes, whereas preoperatively BCVA $>6/60$ was present in 13 (56.5%) eyes and $>6/18$ in 4 (17.4%) eyes. Visual acuity of case 8 with corneal decompensation did not improve after surgery due to persisting edema. BCVA of both eyes of case 7 improved to 6/9 although the acceptance with the previous correction of -32 DS OD and -33 DS OS was 6/18 in both eyes and this was attributable to minification factor of thick concave lenses.

Management of glaucoma

Five patients presented with secondary glaucoma. Two eyes of two patients presented with grade 2 corneal edema due to glaucoma. Six of the ten eyes presenting with glaucoma had IOP of >21 mm Hg, which underwent lensectomy and the IOPs came down to <16 mm Hg. Two of these eyes had the crystalline lens in the anterior chamber with the pupil constricted and open anterior chamber angles and the rest had pupillary block mechanism of glaucoma. In addition to lensectomy, these patients also received preoperative glaucoma medication.

Table 1 Clinical profile of microspherophakia patients

Cases	Eye number	Age/Sex		Presentation BCVA	Presentation IOP (mm Hg)	Family history	Chief complaint	Diagnosis	Refractive error	Anterior segment	CD (mm)	Lens	Posterior segment	Medical treatment	AL	Pre-op Km	WTW	Pre-op specular count (cells/cumm)	CCT (μm)	Surgery	PI	Post-op BCVA	Post-op IOP (mm Hg)	Post-up specular (cells/cu mm)
1	1	7/F	OD	distance FCCF, near NA	26 on 1/2 Tab Acetazolamide TDS, ED Brimonidine +Timolol BD	none	DOV BE, pain, redness	BE microspherophakia with RE anteriorly dislocated lens LE superonasally subluxated lens	poor glow, no streak formed	RDS, ciliary congestion, corneal edema+, AC shallow, 270° anterior synechiae	10.5/11	spherophakia with anterior subluxated lens;	re media hazy	Acetazolamide tablets, Syrup Glycerol, BE Homide, Brimonidine	22.5	41.0/44.0 @ 109°	11.8	3231	543	intralenticular lens aspiration, Left aphakic	yes	FCCF–3/60 with +10D	11	3127
	2		OS	distance 6/36 – NF; near N18 improving to N8 with -14D	15 on 1/2 Tab Acetazolamide TDS, ED Brimonidine +Timolol BD				aphakic +18DS/-2DC @ 180°; phakic - 14DS/-6DC @ 180°. Improving to 6/24 with -16 DS/-2DC @ 180°	AC shallow, subluxated lens	10.5/11	spherophakia with superonasally subluxated lens	le CDR-0.3:1, NRR healthy, FR sharp		22.48	43.0/45.25 @ 13°	11.9	3426	552		yes		4	
2	3	8/M	OD	distance 1/60; near >N36	9	none	DOV BE, pain, redness, watering RE	BE microspherophakia with RE anteriorly dislocated lens LE inferiorly subluxated lens	streak -20 vert/-18 hori;	ciliary congestion, AC shallow irregular, inferonasal PAS, 8 mm fixed dilated pupil with sphincter tears	10/10.5	anteriorly dislocated microspherophakia with broken zonules	re CDR-0.6:1, temporal pallor+		22.87	45.0/46.0 @ 170/80;	11.6	3127	567	intralenticular lens aspiration with ACIOL	yes	6/60	12	3097
	4		OS	1/60, >N36	14				streak -22 vert/-22 hori;	inferiorly subluxated lens	11/10.5	clear crystalline microspherophakia with phakodonesis with inferior subluxation	le CDR-0.6:1, temporal pallor		22.73	44.75/45.50 @ 6/96	11.3	3234	561	intralenticular lens aspiration , ACIOL	yes	6/36	16	3136
3	5	7/F	OD	distance FCCF–NFI near <N36	25 on Acetazolamide 1 tab TDS and ED Brimonidine +Timolol BD BE	none	DOV BE (R>L) x 3 years	BE microspherophakia with secondary glaucoma	-14DS acceptance 4/60; ; streak phakic -26D vert/-24D hori, ; aphakic +16D vert/+14D hori,	lens in anterior chamber	12.6/12.4	microspherophakia with anterior subluxation with broken zonules; lens in anterior chamber	re CDR-0.9:1, NRR thin	Tab Acetazolamide 1/2 TDS, BE Timolol, Brimonidine, Homide, Dorzolamide drops	21.97	36.49/40 @ 77/167	13.9	3324	535	lens aspiration and left aphakic	yes	FCCF–5/60 with + 18D; near N8 with + 20D,	4	3269
	6		OS	6/60–6/36P; near N36	24 on Acetazolamide 1 tab TDS and ED Brimonidine +Timolol BD BE				-14DS acceptance 6/36; streak phakic -12D vert/-14D hori; aphakic +18D vert/+14D hori		13.1/12.9	microspherophakia with inferior subluxation with stretched zonules	le CDR-0.9:1, NRR thin		21.68	38.66/40.56 @ 116/26	14.2	3259	557	lens aspiration and left aphakic	yes	1/60–6/36 with +18D; near N8 with + 20D	4	3201
4	7	23/M	OD	2/60–6/36P; near N36–N18 with +3DS	8	positive family history in elder brother	sudden DOV, gradually progressing in RE x 1 month	Marfan's syndrome with RE spherophakia with BE Ectopia lentis	streak -8.5 vert/-9 hori; acceptance with -10 DS/-0.5 DC @ 90° is 6/12P	inferiorly subluxated lens	12/12	anteriorly subluxated lens with spherophakia with inferonasal 9 clock hours subluxation with zonular breaks	cdr 0.4:1, NRR pale FR dull		20.99	43.00/43.50 @ 20°	12.5	1012	523	intralenticular lens aspiration with ACIOL	yes	HMCF on day 1; 6/12P on day 4	12	961
	8		OS	6/18–6/9P; near n6	10				streak -1 vert/-1.5 hori; acceptance with -2.5 DS/-0.5 DC @ 90° –6/9P	inferiorly subluxated lens	12/12	nasally subluxated lens with 6 clock hours subluxation with vitreous with zonular breaks	cdr 0.4:1, NRR pale FR dull		20.54	42.0/43.5 @29°	12.3	3507	532	intralenticular lens aspiration with ACIOL	yes	FCCF on day 1; 6/18 on day 4	15	3422
5	9	20/F	OD	6/18–6/12P; near N10	14	positive family history in elder brother	DOV BE, painless 7-8 years	BE microspherophakia with subluxated lens	streak -7.0 vert/-8 hori; acceptance with -9 DS/-4 DC @ 180° - 6/12P	nasal ACD< Temporal ACD; no PAS/ shallow angle; Iridodonesis	11.5/12	spherophakia with superonasal subluxation 9 clock hours, pigments on zonules, phakodonesis	CDR 0.4:1, NRR healthy, FR sharp		23.17	41.5/43.75 @ 4°	12.5	3277	519		yes		17	
	10		OS	3/60–6/36P; N12	12				streak -12.0 vert/-11 hori; acceptance with -12 DS/-1 DC @ 180° -6/36	nasal ACD< Temporal ACD; no PAS/ shallow angle; Iridodonesis	12/12	spherophakia with superonasal subluxation 9 clock hours, pigments on zonules, phacodonesis	CDR 0.4:1, NRR healthy, FR sharp		24.99	41.5/43.5 @ 15°	12.49	3427	547	lens aspiration with CTR placement with PCIOL in bag	yes	6/9	12	3289
6	11	10/F	OD	2/60 – 6/60 naked eye; with gl 6/18–NFI, near N18–N10 with + 16D; 1 month back BCVA 6/36	24	none	sudden DOV BE x 4 years, dilation of LE pupil since 1 month and pain	BE microspherophakia with secondary glaucoma	streak aphakic +14D @ 145°/+13D @ 35°; phakic - 16D vert/-14D hori; aphakic + 12.5 DS/-1 DC @ 145° acceptance – 6/12; NV N18–N10 with +16D	intermittent divergent squint; iridodonesis	11.5/11.5	superonasal subluxated crystalline lens, phacodonesis, broken zonules	CDR 0.3:1, NRR healthy, FR sharp	Eye drops Timolol bd	23.42	45.0/46.0 @ 20/110	12	3986	502	intralenticular lens aspiration with ACIOL	yes	6/24	10	3768
	12		OS	2/60–3/60 naked eye; with gl 6/24–NFI, near N10–NFI	22				streak phakic -16D vert/-7D hori; acceptance with -7DS – 6/18; no acceptance of cylinder; NV - N10 with -7D	ciliary congestion, corneal edema,lens present in anterior chamber, iridodonesis	11/11	anteriorly dislocated crystalline lens with microspherophakia, phacodonesis, broken zonules	CDR 0.3:1, NRR healthy, FR sharp	Eye drops Timolol bd	23.65	43.75/44.50 @ 10/100	11.3	2616	506	intralenticular lens aspiration with ACIOL	yes	6/12	14	2519
7	13	12/F	OD	distance 1/60 – 3/60 naked eye; with gls 6/18; near N8	14	positive family history in elder brother, brother operated for bilateral RD	DOV BE, painless 10 years	BE microspherophakia	streak -36D vert/ -34.5D vert; acceptance with -37.5 DS/ +1.5DC @ 90 -6/18	AC centrally shallow, iridodonesis	11.5/11.5	phacodonesis with microspherophakia	CDR 0.3:1, NRR healthy, FR sharp		26.38	42.25/43.50 @ 40/130	12.3	2473	538	lens aspiration + ACIOL	yes	6/18	11	2329

Table 1 (continued).

	14		O S	distance 1/60–6/60 naked eye; with gls 6/18; near N8	16				streak -36D vert/ -35D vert; acceptance with -37 DS/+1 DC @ 180 - 6/18	AC centrally shallow, iridodonesis	11.5/ 11.5	phacodonesis with microspherophakia	CDR 0.3:1, NRR healthy, FR sharp		26.99	42.0/4 34.0 @ 160/7 0	12.5	2372	529	lens aspiration + ACIOL	yes	6/9	12	2184
8	15	32/F	O D	distance FCCF; near <N36	13	none	DOV RE>>LE x 1 month; redness, pain	BE microspherophakia with RE anteriorly dislocated lens with corneal decompensation LE nasally subluxated lens	poor streak formation; subjective refraction with +14D – no acceptance; subjective refraction with +16D – 1/60	mild corneal edema in center, irregular AC shallower nasally	11.5/ 11.5	spherophakia with anteriorly dislocated crystalline lens	CDR 0.2:1, NRR healthy, FR sharp		24.38	39.0/4 1.0 @ 180/9 0	11.4	3189	520	intralenticular lens aspiration + ACIOL	yes	1/60 – NFI	16	3026
	16		O S	distance 4/60 – NFI; near N10	12				streak phakic -16D vert/ -14D hori; acceptance with -16 DS/ -2 DC @ 180' is 4/60	irregular AC shallower nasally	11.5/ 12	spherophakia with nasally dislocated crystalline lens from 7-11 o'clock	CDR 0.2:1, NRR healthy, FR sharp		22.62	40.25/ 41.5 @ 150/6 0	11.8	3258	511		yes		16	
9	17	40/M	O D	distance 6/36, N10	18	none	DOV BE x 6 years	BE microspherophakia with inferotemporal subluxated lens with secondary glaucoma	-8.50 DS/-8.0 DC @ 157'	AC shallow	11.7/ 11.5	spherophakia with inferior subluxation	CDR 0.8:1, NRR inferiorly thin, FR sharp	Eye drops Latanoprost HS	20.93	41.75/ 47.5 @ 150/6 0	11.7	2175	550	intralenticular lens aspiration with trabeculectomy, left aphakic	yes	FCCF, N24	10	1978
	18		O S	distance 6/24, N8	24				-10.0 DS	AC shallow	13.0/ 12.8	spherophakia with inferior subluxation	CDR 0.9:1, NRR inferiorly thin, FR sharp	Eye drops Latanoprost HS	20.91	43.0/4 4.75 @ 165/7 5	13.7	2204	603	intralenticular lens aspiration with trabeculectomy, left aphakic	yes	HMCF, <N36	18	1923
10	19	2/M	O D	Cardiffs 50 cm –6/60	Tonopen 12	none	abnormal movements of BE	BE microspherophakia with megalocornea	-22 DS/-6.5 DC at 180	Megalocornea, AC shallow	13/14	globular lens behind pupil	CDR 0.2:1, NRR healthy, FR sharp		21.4	42.25/ 46.25 @ 25/11 5	13.5	2100	604	lens aspiration with CTR in bag, PCIOL in bag	yes	follows light	8	2021
	20		O S	Cardiffs 50 cm –6/60	Tonopen 16				-22 DS/-7 DC @ 180'	Megalocornea, AC shallow	13/14	globular lens behind pupil	CDR 0.2:1, NRR healthy, FR sharp		20.9	42.50/ 45.75 @ 157/6 7	13.7	2276	591	les aspiration with CTR in bag, PCIOL in bag	yes	follows light	10	2088
11	21	31/M	O D	distance 6/36, near N12	16	positive family history in sister	DOV BE, painless, 1 year	BE microspherophakia with posteriorly dislocated cataractous lens in anterior vitreous	+6 DS/-2DC @ 40' acceptance - 6/24	Pigments over endothelium, Deep AC, iridodonesis, ruff atrophy	11.8/ 12.3	posteriorly dislocated cataractous lens in inferior vitreous	CDR 0.3:1, NRR healthy, FR sharp		27.1	43.0/4 4.5 @ 80/17 0	12.5	2187	564	PPV with phacofrag with SFIOL	no	1/60	6	1986
	22		O S	distance 6/60, near N12	15				+6 DS/-2 DC @ 30' acceptance - 6/36	Pigments over endothelium, Deep AC, iridodonesis, ruff atrophy	12.5/ 12.5	posteriorly dislocated cataractous lens in anterior vitreous	CDR 0.3:1, NRR healthy, FR sharp		27.24	43.0/4 4.0 @ 83/17 3	12.1	2246	536	PPV with phacofrag with SFIOL	no	FCCF	4	2049
12	23	24/M	O D	distance 6/60, near <N36	13	none	DOV BE 10 years, pain LE since 3 months	Marfan's syndrome with BE microspherophakia with secondary glaucoma	-14.5 DS/ -1 DC @ 180' acceptance - 6/24	AC irregular	12.5/ 13	spherophakia with inferonasal subluxation	CDR 0.5:1, NRR healthy, FR sharp	Eye drops Latanoprost HS	21.93	42.0/4 4.5 @ 93/3	12.5	2614	561	intralenticular lens aspiration with ACIOL	yes	6/18	6	2520
	24		O S	distance 1/60; near <N36	14				-9.5 DS/ -4 DC @ 180 acceptance - 6/36	AC irregular, central corneal edema	12.5/ 13	spherophakia with inferonasal subluxation	CDR 0.5:1, NRR healthy, FR sharp	Eye drops Latanoprost HS	21.69	46.0/4 7.0 @ 130/4 0	12.5	1719	547	intralenticular lens aspiration with ACIOL	yes	6/18	6	1639
13	25	6/M	O D	distance 6/60, near N8	12 mm Hg	none	DOV BE x 3 years	BE microspherophakia	acceptance with -14 DS - 6/60	corneal haze, AC shallow	12/11 .5	Spherophakia with shallow AC	CDR 0.3:1, NRR healthy, FR sharp		20.01	43.0/4 2.75@ 30/12 0	12.1	2320	532	lens aspiration and left aphakic	yes	6/18	10	2196
	26		O S	distance 5/60, near N8	14				acceptance with -16.5 DS - 5/60	corneal haze, AC shallow	12/11 .5	Spherophakia with shallow AC	CDR 0.3:1, NRR healthy, FR sharp		20.86	42.5/4 3.0 @ 90/0	11.8	2187	519	lens aspiration and left aphakic	yes	6/12	12	2064

Table 2 Ocular parameters of sample population

Clinical parameters	*Mean ± SD*	
IOP at presentation ($n=26$)	15.46 ± 4.89	
Preoperative IOP of operated eyes ($n=23$)	16.13 ± 5.41	
Postoperative IOP of operated eyes ($n=23$)	10.39 ± 4.02	*P*-value <0.001
Preoperative endothelial cell count of operated eyes ($n=21$)	2633.35 ± 668.52	
Postoperative endothelial cell count of operated eyes ($n=21$)	2455.64 ± 652.36	*P*-value = 0.36
Preoperative CCT ($n=24$)	544.19 ±27.09	

Two patients had to be started on oral carbonic anhydrase inhibitors apart from topical multi-drug therapy and three patients could be managed with monotherapy. Nineteen eyes had normal optic nerve head (ONH), whereas six eyes showed increased ONH cupping and glaucomatous changes with high IOP at presentation. Fundus for one eye could not be evaluated in view of corneal edema and media haze. Eyes that had an ACIOL implantation all underwent PI. Fellow eyes with spherophakia, which had been left for observation also underwent a PI. Two eyes of one patient (case 9) showed elevated pressures and advanced glaucomatous cupping at presentation for which a combined lensectomy with trabeculectomy was performed and on follow-up he continuously showed elevated pressures (>40 mm Hg) for which bleb revision surgery had to be done in both eyes following which the IOPs came down to <10 mm Hg. Case 3 presented with raised IOP >30 mm Hg in the right eye at 3 months' follow-up after lensectomy on multiple drug therapy and underwent trabeculectomy in right eye following which the IOP came down to <10 mm Hg.

Systemic associations

Four patients (cases 4, 5, 7, and 11) had positive family history of spherophakia in their siblings (elder brothers in 4, 5, and 7, sister in 11), of whom two patients (cases 4 and 11) were known cases of Marfan's syndrome with arachnodactyly and high-arched palate. Both these cases had aortic root dilatation. One of these three elder brothers had a history of spherophakia with high myopia and bilateral retinal detachments for which 25G vitreoretinal surgery with silicone oil injection had been performed. One patient (case 2) had a history of seizures since 3 years of age with a right-sided hemiparesis and was found to have raised urine and serum homocysteine levels diagnostic of homocystinuria; however, no family history was found. This child was referred to the Paediatrics department for further management. The rest patients were believed to be having isolated spherophakia.

No intervention

No surgical intervention was performed on three eyes, of which one eye had angle closure due to pupillary block and was kept on glaucoma medication. PI was performed for all of these eyes.

Discussion

Most spherophakia patients present in adolescence or early adulthood and patients having systemic associations may present earlier, the median age in our study being 12 years. Most of our patients had a history of insidious onset diminution of vision and pain in some of the cases of secondary glaucoma, with diminution of vision being the commonest complaint. The condition may be isolated, familial, or associated with systemic disorders like homocysteinemia, Weil–Marchesani syndrome, Marfan's syndrome, Alport's syndrome, hyperlysinemia, megalocornea, spherophakia-secondary glaucoma, arrhythmogenic right ventricular dysplasia type 1, and so on.[2] The spherical lens may dislocate superotemporally, inferonasally, or inferiorly. The intraocular findings are a spherical lens, iridodonesis, and axial myopia, and the physical features depend on the syndrome associated, like arachnodactyly/brachydactyly, increased upper segment–lower segment ratios, tall/short stature, high-arched palate, joint stiffness, and congenital heart defects. Homocysteine levels may be raised in the urine and serum. Patients may also have musculoskeletal or metabolic diseases. But patients usually do not have buphthalmos, enlarged cornea, abnormal angle structures, or increased axial length associated, which differentiates the condition from primary congenital glaucoma. Although most of our cases were isolated spherophakia with inferior subluxation, we have described one case of homocystinuria with a history of seizures and hemiparesis and two cases of Marfan's syndrome. Cases of homocystinuria have been found to be associated with seizures and hemiparesis due to a hypercoagulable state.[6] Cardiovascular associations are present in virtually most adults with Marfan's.[7] Both of

our cases had aortic root dilatation associated but ECG was normal in all of them.

Lens extraction/lensectomy

Lensectomy has been previously described as an option for managing the dislocated lens,[8] following which visual rehabilitation becomes important, with probable options being aphakic spectacles, contact lenses, and IOLs. The choice of the IOL depends largely on the surgeon and patient factors. Angle-supported anterior chamber lenses (ACIOL; Figure 2c) and iris-enclavated lenses[9] are commonly used options. Posterior chamber IOL (PCIOL) with/without CTRs/segments (CTS)[10] and scleral-fixated IOL (SFIOL)[11] have also been described in some isolated case reports. After IOL implantation patients need to be followed for long term to observe for amblyopia and occlusion therapy. Despite all efforts, amblyopia becomes difficult to manage, with compliance of glasses usage being poor in children, and ametropic amblyopia has been reported in 50% of patients with familial lens subluxation.[12]

Angle-supported ACIOLs have been reported to be associated with corneal endothelial cell loss, peripheral anterior synechiae (PAS) formation and glaucoma due to chronic anterior chamber irritation and modern users may have become skeptical of its use in younger population.[13] Dehgan *et al*[14] used angle-supported ACIOLs in most of their aphakic patients combined with a PI, with only few cases of displacement, IOL capture, and endothelial touch, which only reaffirms the safety of a properly sized ACIOL surgery done in experienced hands.[15,16] The angle-supported ACIOL when planned rather than implanted in a complicated case has better results. Iris-supported ACIOLs is also an option for hereditary lens subluxation cases because of larger WTW in Marfan's cases posing difficulty in choosing angle-supported IOL size. Iris-enclavated lenses may be placed anterior[9] or posterior[17] to the iris. But these iris claw lenses are hinging on a light sensitive mobile structure. Although ACIOL entry requires a large corneal incision which may later lead to astigmatism, it does have the advantage of better pupillary mobility compared to iris claw lens. (Supplementary Video 2)

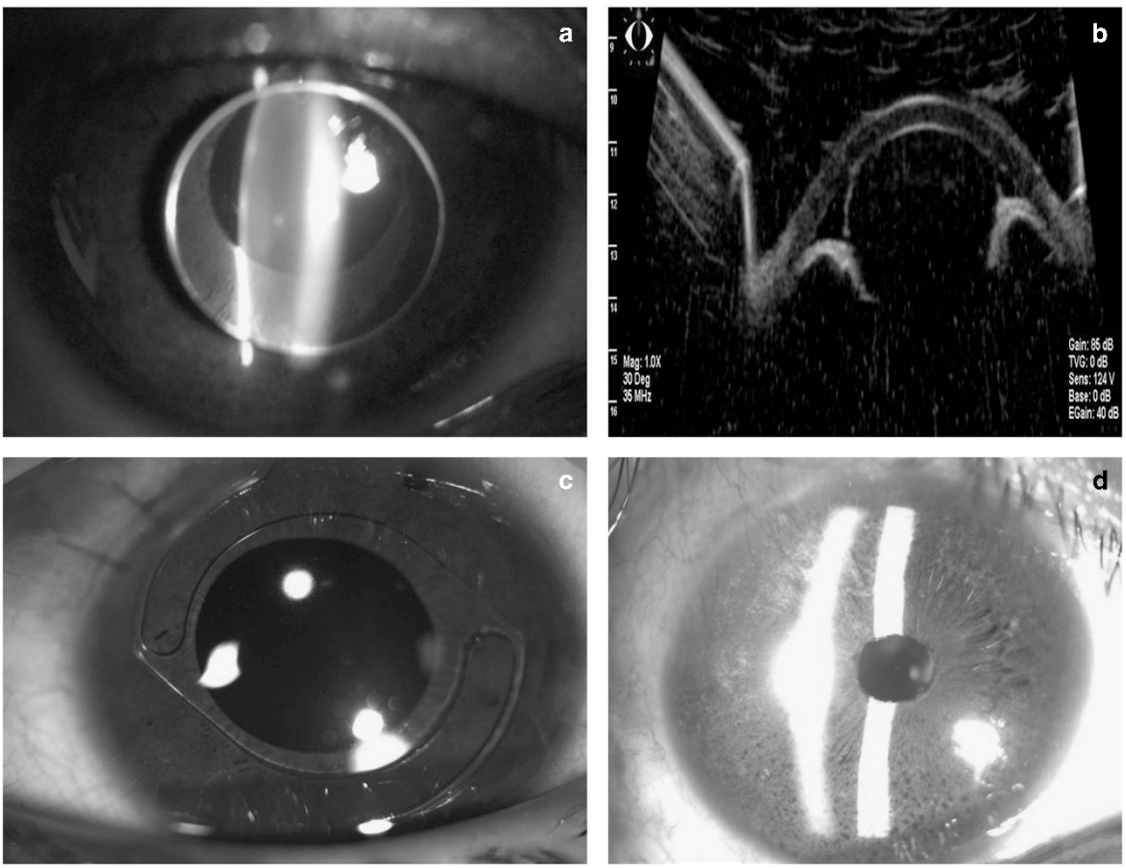

Figure 2 (a) Anteriorly dislocated spherophakic lens. (b) UBM showing lens in anterior chamber touching the cornea. (c) ACIOL in spherophakic patient. (d) SFIOL in spherophakic patient. A full colour version of this figure is available at the *Eye* journal online.

Recently iris-supported ACIOLs have been used in pediatric aphakics with good visual outcome and no significant effect on the endothelium.[18] Iris-supported ACIOLs seem to be a better option for hereditary lens subluxation cases because of larger WTW in Marfan's cases posing difficulty in choosing angle-supported IOL size. Iris-enclavated lenses may be placed anterior[9] or posterior to the iris. Some studies have demonstrated that although immediate postoperatively ECDs may be affected, reported to be 1.5–10% at 3 years,[19] they stabilize few years after. Although traumatic de-enclavations have been rarely reported till date, they are a cause for concern with these lenses[20] specially in children. In spite of the popularity of Artisan iris-enclavated lenses, considering their inflated prices and easy local availability and affordability of angle-supported lenses, we opted for the latter in our study.

PCIOL placement is controversial as the zonules are developmentally weak and there is a possibility of the bag lens complex falling into the vitreous.[21] Khokhar *et al*[22] have described a 'dual-support technique' of insertion of a CTS with CTR in a case of spherophakia and have opined that this may help overcome zonular weakness. Also, Cionni (M-CTR) fixation to the sclera with 9-0 polypropylene (prolene) has been performed. A CTR has been said to strengthen the bag and reduce the risk of IOL subluxation.

SFIOL has been documented as a viable option for aphakic children (Figure 2d) and has been said to be rid of all these angle-related controversies, however, it requires an effective vitrectomy and proper scleral tunnel fixation. However, there has been a concern regarding SFIOL use in pediatric patients because of elastic and less rigid sclera compared to adults. SFIOL haptics have to be buried under the scleral flaps with polypropylene sutures and reports have shown that due to lack of fibrosis around lens loops, the suture is the only support for the lens.[23] Transscleral suture exposure has been reported at 14.7–17.9%.[23] Although prolene is theoretically non-absorbable, there is a possibility of late decentration caused by suture degradation even years after, and this is particularly bothersome for the pediatric patient.[24] Reports of suture breakage in SFIOLs further substantiate the concern.[25] To summarize, SFIOL is a difficult procedure technically and may be associated with reported complications.[13,26] In cases with a WTW >13 mm such as megalocornea, SFIOL, or ACIOL are not preferred due to possible complications of IOL instability and decentration, respectively. Iris claw lens enclavated within the iris stroma may be more appropriate in such cases, with good visual outcome and no significant effect on the endothelium.[18]

None of our ACIOL patients presented with corneal decompensation. Likewise, PCIOL with M-CTR in the bag was stable and SFIOL patients did not present with IOL tilt or haptic extrusion till the last follow-up. The lenticular myopia of the order of -10 to -15 D is usually the first ophthalmologic finding in any spherophakia patient with usually normal axial lengths and corneal topography,[27] which can considerably affect the quality of life. Hence, clear lens extraction with proper IOL placement can be considered as a method of correcting the unacceptably high myopia. In the study, visual improvement of $>6/18$ Snellen's was reported in 43.5% and $>6/60$ in 78% of our operated eyes. We believe that in this study we have reported the largest number of cases managed by lens extraction followed by different techniques of visual rehabilitation using mostly angle-supported ACIOL and few ones with PCIOL and SFIOL. The easy availability and affordability of angle-supported lenses makes it more viable compared to iris fixated and SFIOL specially in developing countries.

Spherophakia may be associated with angle-closure glaucoma, which may be because of pupillary block due to the spontaneous shift of iris–lens diaphragm anteriorly[4] or the lens itself dislocating into the anterior chamber (Figure 2a) to cause angle block mechanically. The latter phenomenon has been labeled as inverse glaucoma[28] and not unlike malignant glaucoma, this condition is deteriorated by miotics and relieved by mydriatic agents.[29] Spontaneous dislocation has been described as a common cause of lens-induced glaucoma.[30] Post-traumatic dislocation of the lens may also occur due to weak zonules. The migration of the lens into the anterior chamber may be intermittent causing acute glaucoma crises every time[5] and may eventually lead to retinal detachment due to the vitreous traction. A chronic pupillary block due to the forward migration of iris–lens diaphragm may result in crowding of the trabeculae (Figure 2b) and unrelieved, may lead to PAS formation and trabecular damage due to raised IOP.[31] The lens may also dislocate posteriorly into the vitreous cavity[4] as was found in one of our cases. Earlier concepts by Willi *et al*[32] involved performing a laser PI to relieve pupillary block in such eyes and a surgical PI in case it failed.[33] However, Asaoka *et al*[34] suggested that trabeculectomy was necessary to control the IOP. Another school of thought by Willoughby *et al*[35] had developed, which stated that lensectomy would be able to control glaucoma in spherophakia, although Yasar[36] suggested that it was a temporary option requiring a trabeculectomy subsequently for IOP control. Most of the eyes presenting with raised IOP in our study were managed with lensectomy with PI, which led to a significant lowered IOP ($P<0.001$) in all of them and normal IOP on follow-up. One patient underwent combined lensectomy with glaucoma surgery followed by bleb revision, whereas another patient who presented with inverse glaucoma

showed uncontrolled IOP post lens extraction and a trabeculectomy was performed for IOP control.

In summary, spherophakia is a disabling disease, which presents bilaterally with high myopia along with a risk of long-term glaucoma and systemic associations if it is a part of a syndromic manifestation. This can be tackled by lens extraction and IOL placement for visual rehabilitation and glaucoma treatment. Glaucoma management may be refractory and require further surgeries but overall the outcomes are good. The ACIOL are still a viable option when planned and put under experienced surgical hands.

Summary

What was known before

- Spherophakia presents with varying degree of lenticular myopia, glaucoma, and subluxation of the crystalline lens.
- Lensectomy and trabeculectomy have been described for management.

What this study adds

- Lensectomy alone along with ACIOL and SFIOL is a viable option of controlling glaucoma and correcting high lenticular myopia.

Conflict of interest

The authors declare no conflict of interest.

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Supplementary Information accompanies this paper on Eye website (http://www.nature.com/eye)